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Dermatofibrosarcoma Protuberans on the Chest with a Variety of Clinical Features Masquerading as a Keloid: Is the Disease Really Protuberant?

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Dear Editor:

Dermatofibrosarcoma protuberans (DFSP) is regarded a locally invasive, low-grade sarcoma. DFSP is typically characterized by its protuberant appearance. The DFSP in our patient had a clinically heterogeneous appearance, which is very rare, so the DFSP was initially misdiagnosed as a keloid.

A 46-year-old male presented with a progressing skin lesion on his anterior chest. The erythema with itching had developed in his 20s, and the nodules appeared gradually. The nodules were thought to be keloids, so he received intralesional steroid injections for a few years at a dermatological clinic. However, that therapy had little effect, and he was referred to our department. He presented with an 8.0×10.0-cm shiny, atrophic skin-colored plaque, in which indurated erythema and painful keloid-like nodules were observed (Fig. 1). These nodules were also found on his chest area. The atrophic plaque (Fig. 2A), erythema (Fig. 2B), and nodule (Fig. 2C) were biopsied.

Histologically, the 3 samples showed common basic structures. The epidermis consisted of atrophic and spindle-shaped cells, showing little atypism; the cells proliferated in a storiform pattern throughout the dermis and even extended to the lobular structures of fatty tissue. The tumor cells were CD34 positive.

To identify the histological differences according to clinical status, we examined the cellularity and positive staining rates for Mib-1 in the atrophic plaque, erythema, and nodule. The cellularity count for each sample was performed in 10 randomly selected areas at high magnification (×200). The degree of cellularity in the nodular lesion (Fig. 2F) was higher than that in the other lesions

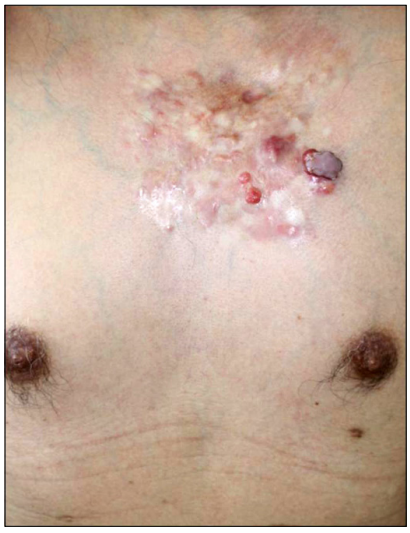

Fig. 1. Clinical features of the 3 different lesions: shiny atrophic plaque (8.0×10.0 cm), indurated erythema, and painful, keloid-like red nodules (largest nodule: 2.0×1.5 cm) on the anterior chest.

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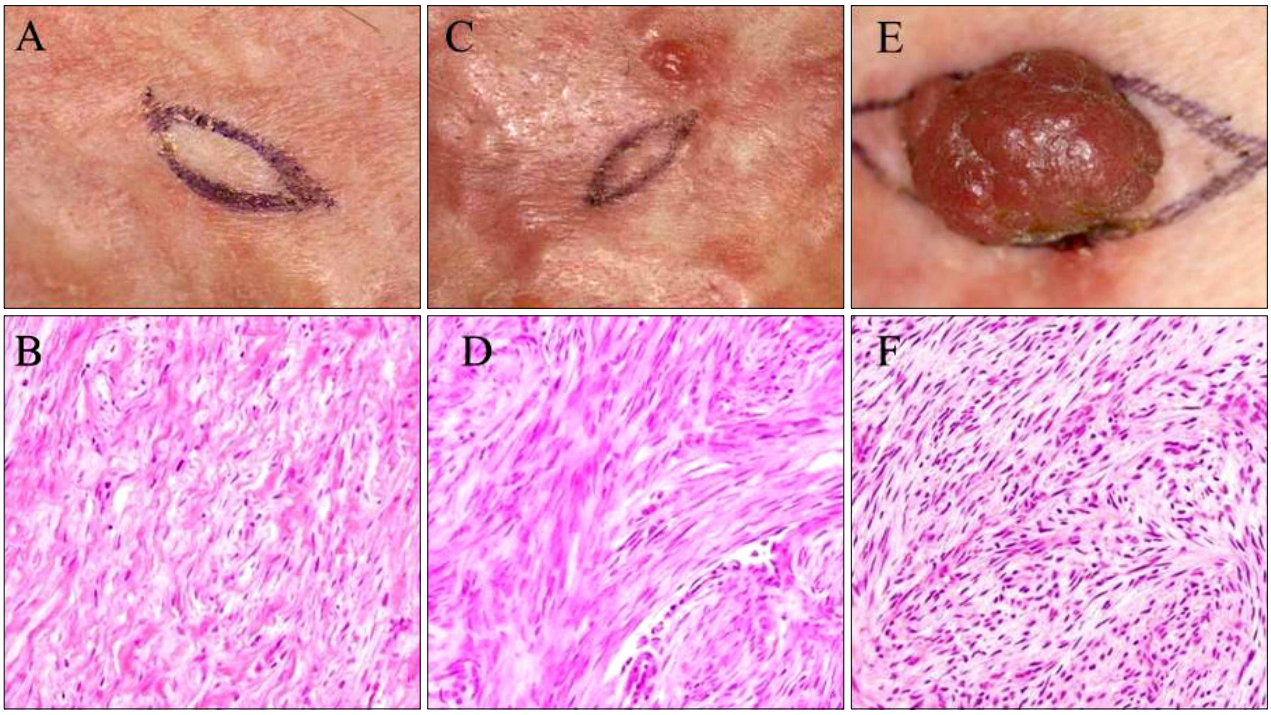

Fig. 2. Histopathological features of the lesions (H&E, ×200). Atrophic plaque (A); a few proliferating spindle cells are seen (B). Erythematous plaque (C); storiform pattern is evident, but cellularity is low (D). Keloid-like nodule (E); typical pathological findings of DFSP; the cells show a storiform pattern, which is the dominant pattern, and infiltration into the subcutis. Cellularity is the highest in this lesion (F).

(Fig. 2D, E). The mitotic index was similar among the 3 lesions.

Later, the tumor was resected along the planned 3-cm margin. The overlying skin defect was reconstructed using a flap from the greater pectoral muscle. At the follow-up after 2 years, the patient was found to be free of the disease.

DFSP is typically characterized by its protuberant appearance during early or middle adulthood. However, several cases of clinical and pathological unusual variants, such as Bednar's tumor and myxoid, sclerosing, fibrosarcomatous, atrophic, and nonprotuberant DFSP have been reported[1]. Our patient showed unique and various clinical features in a single location, including atrophic plaque, indurated erythema, and keloid-like nodules. What factors cause these different clinical appearances?

The atrophic variant of DFSP is most commonly found on the trunk of women[2]. Furthermore, atrophic DFSP is more common among children and young adults[3] than among the elderly. Martin et al.[4] reported a nonprotuberant form of DFSP, and nearly half of their patients identified their early DFSP-related skin changes as patches, and the nonprotuberant stage lasted for 7.6 years. In those cases, the clinical appearance of the lesions resembled that of morphea, morpheaform basal cell carcinoma, atrophoderma, or angioma lesions. The *COL1A1-PDGFB* fusion gene is present in all the cases of DFSP subtypes. The different types of fusions between *COL1A1* and *PDGFB*, however, are not related to the differences in clinical or histological features[5]; the factors contributing to these clinical differences have not yet been clarified.

DFSP is not always protuberant, and awareness of this rare clinical presentation may assist in the early diagnosis of unusual variants of DFSP.

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Toxic Epidermal Necrolysis in a Patient with HLA-B*5901 Haplotype Caused by Topical and Oral Carbonic Anhydrase Inhibitors

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Dear Editor:

Toxic epidermal necrolysis (TEN) and Stevens－Johnson syndrome (SJS) are severe life-threatening mucocutaneous diseases that involve different extents of epidermal detachment and severity[1]. Carbonic anhydrase inhibitors (CAIs) are used to decrease intraocular pressure (IOP) in glaucoma[2] and may rarely induce TEN/SJS. Moreover, strong genetic associations between human leukocyte antigen (HLA)-B*5901HLA alleles and methazolamide-induced TEN/SJS have been discovered in Koreans[3]. Herein, we report a case of TEN in a Korean woman with HLA-B* 5901 haplotype, who had taken methazolamide for 1 day and instilled brinzolamide for 10 days.

An otherwise healthy 33-year-old Korean woman presented with pruritic, multiple, coalescent, erythematous patches with vesicles on the face and trunk involving erosive oral mucosa (Fig. 1A). Twenty days prior, she visited an ophthalmic clinic complaining of blurred vision after taking phendimetrazine, an anorectic agent, for 2 days. Her IOP was elevated and was treated with methazolamide, brinzolamide 1%/timolol 0.5%, brimonidine 0.15%, and prednisolone 1% eye drops for 1 day. Nevertheless, her IOP remained elevated, and both eyes were treated with argon laser iridotomy. Brinzolamide 1%/timolol 0.5% and brimonidine 0.15% eye drops were administrated for an additional 9 days. Two days after stopping the eye drops, she noted pruritic erythematous macules on the face. Her cutaneous lesions were aggravated and expanded distally with conjunctival and oral mucosal involvements over several days. She had no history of drug allergies, including sulfonamide antibiotics. Personal and family histories were unremarkable. Routine laboratory test results were normal. HLA typing showed B*5901 and B*5204. She was initially treated with dexamethasone 10 mg daily for 2 days, but epidermal necrolysis worsened and became confluent (Fig. 1B, C). Intravenous immunoglobulin (3.5 g/day) was infused for 4 days. Her skin lesions gradually improved

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